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CASE REPORT

Coronary artery fistula; non-invasive diagnosis by cardiovascular magnetic resonance imaging

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Coronary arteriovenous fistulas are among rare anomalies of coronary arteries. Role of X-ray angiography is well established in identification and characterization of these anomalies, however there accurate course and termination is often not defined. We demonstrate role of routine cardiovascular MRI in non-invasively diagnosing and characterizing the course and termination of these anomalous coronary branches.

Key Words: Coronary arteriovenous fistula; Magnetic resonance imaging; Non invasive

1. Introduction

Coronary arteriovenous fistula (CAF) is considered infrequently in the differential diagnosis of left to right shunt, myocardial ischemia, or congestive heart failure because of relatively uncommon occurrence. They are more commonly congenital (0.2 to 2.5% angiographically) than acquired after coronary intervention, thoracic trauma, endomyocardial pacer lead placement, or endomyocardial biopsy. Sakakibara et al. (1) described two types of coronary arteriovenous fistula: type A called proximal or side to side fistula and type B called distal or end artery type fistula also known as cameral fistula. The majority of coronary fistula drains into low pressure right sided structure and present with clinical constellation of dyspnea on exertion, angina, congestive heart failure symptoms, and, rarely, myocardial infarction from coronary steal phenomenon. Rarely, patients may present with endocarditis or embolization from the thrombotic material at the mouth of the fistula. Here we describe two cases of coronary arteriovenous fistula non-invasively diagnosed by cardiovascular magnetic resonance imaging (CMR) after initial ambiguity by x-ray angiography.

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2. Case 1

A 37 year old Caucasian male with remote history of arteriovenous malformation (AVM) removed from his left arm and chronic AVM in his left eye presented with chest tightness radiating to the left side of his chest. The resting ECG was normal, and a stress test revealed moderate inferoseptal, inferior and inferolateral defect with moderate to marked reversibility noted in inferolateral area. Coronary catheterization demonstrated aberrant vessels arising from proximal left anterior descending artery (LAD) with unknown destination despite multiple attempts (Fig. 1A). To further clarify the anatomy, an ECG gated CMR was performed on CVi/Signa 1.5 T GE scanner (Milwaukee, WI). Indeed, three CAF were identified using a combination of non-breathhold and breathhold spin echo and fast imaging employing steady state acquisition (FIESTA) cine images in multiple oblique views (Fig. 1B). Additionally, phase velocity mapping was performed to quantitate the degree of shunt. Two of these aberrant vessels were seen originating from the very proximal LAD coronary artery and traveling anterolaterally, cranially, and finally entering into the main pulmonary artery (MPA). The first vessel entered at 1.7 cm and the second vessel entered more superiorly at 1.9 cm from the anterior pulmonic valve leaflet. A third smaller aberrant vessel originated from proximal left main trunk and entered the pulmonary artery at the level of second most superior vessel (Fig. 1C). The proximal portion of the main pulmonary artery was top normal size (31 mm) while the left and right pulmonary arteries were mildly dilated at 24 mm and 25 mm, respectively, suggesting a high flow state. The right ventricle was moderately dilated (end diastolic dimension = 47 mm) with moderate systolic dysfunction. The left ventricle was mildly dilated (end diastolic dimension = 56 mm) with mild global systolic dysfunction. The LV end diastolic volume was 202 mL, end systolic volume 113 mL, stroke volume 89 mL, and LV EF of 47%. The RV end diastolic volume was 172 mL, end systolic volume 109 mL, stroke volume 63 mL, and RV EF of 37%. The Qp:Qs evaluated by phase velocity mapping was 1.2 and by stroke volume quantification was 1.4. Flow pattern analysis of pulmonary artery suggested 20 to 25% augmentation of normal right ventricular outflow tract flow via AV malformation at the level of MPA. The patient was offered coil closure but declined.

3. Case 2

A 73 year old Caucasian female with past history of unstable angina was referred for CMR after a coronary angiogram suggested the presence of a coronary artery fistula. ECG showed normal sinus rhythm with nonspecific T wave abnormalities in the precordial leads. Coronary arteriography demonstrated non-obstructive CAD and an EF of 70%. A left to right shunt was detected via angiographically identified multiple right coronary artery to pulmonary arterial fistulas.

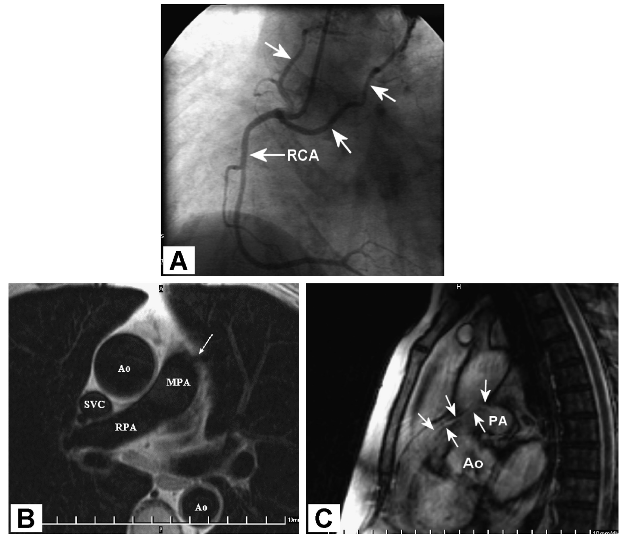

Figure 2. A, B and C are from case 2. 2A) Right coronary angiogram demonstrating two aberrant vessels originating from proximal right coronary artery (RCA) indicated by arrows. 2B) Axial spin echo image, arrow indicate aberrant branch from right coronary terminating into main pulmonary artery. SVC, superior vena cava. 2C) Sagittal FIESTA image demonstrating the aberrant vessel coursing upwards to drain into the pulmonary artery (PA).

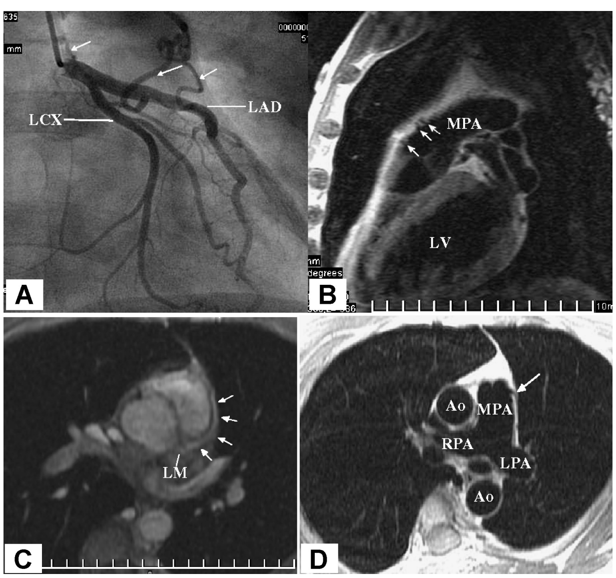

Figure 1. A, B, C and D are from case 1. 1A) Coronary angiogram: Arrows indicate anomalous coronary branches originating from proximal and mid left anterior descending (LAD) coronary artery. LCX, left circumflex coronary artery. 1B) Spin echo sagittal section: Arrows show three terminations of aberrant vessels into main pulmonary artery. 1C) FIESTA axial image: Large aberrant vessel originating from very proximal LAD (arrows). Ao, Aorta; MPA, main pulmonary artery; LM, left main coronary artery; LV, left ventricle. 1D) Axial spin echo image demonstrating the entrance of the aberrant vessel into the main pulmonary artery. LPA, left pulmonary artery; RPA, right pulmonary artery.

The oxygen saturation in the main pulmonary artery was 83%. The left and right pulmonary arteries had oxygen saturations of 69% and 75%, respectively. In an effort to further define these anatomic malformations, CMR was obtained using a GE CVi/ Signa 1.5 T magnet. Using breathhold double inversion recovery sequences (axial and sagittal) and FIESTA (oblique) the coronary tree was visualized. The left main arose from the left coronary cusp and divided into anatomically normal LAD and circumflex arteries. The right coronary arose from the right coronary cusp and immediately divided into a conus branch and a distal branch. Both branches travel anteriorly and superiorly to the left where they entered supravalvular to the pulmonary artery. The first of these entered the MPA anteriorly, 6 mm above the valve plane. The second entered the MPA 18 mm above the valve plane (Fig. 2). A smaller, aberrant, vessel was noted to run confluently with the second branch. The MPA was normal size at 24 mm and the LPA and RPA were also normal size at 17 and 15 mm, respectively. The patient is currently considering coil closure of the fistula.

4. Discussion

The prevalence of CAF is low, and there has been very few studies done as to the appropriate diagnostic modality and management of these patients. The earliest documentation of CAF was by Krause (2) in 1865 and CMR was first used for the diagnosis of CAF by Wertheimer et al. (3) in 1987. Boxer et al. (4) in 1989 used CMR on two pediatric patients to diagnose CAF. Since then very few cases have been described by CMR. We report the first adult cases of CMR phase velocity mapping used to quantitate CAF shunts to our knowledge.

Accurate diagnosis and anatomical relationship of CAF is critical for planning surgical approach and serial follow up. Although there are a number of investigative modalities available very few can characterize the entire course of the CAF incorporating cardiac function evaluation.

Two-D echocardiography has low yield for the origin, accurate epicardial course, and termination of CAF. The diagnosis is improved with transesophageal echocardiography, but it is invasive, requires conscious sedation, and monitoring personnel (5). Computed tomography requires administration of large doses of nephrotoxic radiocontrast, negative chronotropic agent, high x-ray radiation doses, is limited to epicardial vasculature, and complimentary cardiac functional information is lacking. Coronary angiography remains the "gold standard" for the assessment of CAF but is invasive and needs assumptions to the exact course due to the inability to clearly judge the relationship to surrounding structures and termination chamber (6). It is also not suitable for serial follow-ups in asymptomatic patients.

We propose that CMR is ideal alternative noninvasive technique (7, 8). Its inherent 3D imaging volume and large field of view is ideal for anatomical detailing of CAF along with a search for any additional congenital cardiac or vascular associations. Beside exact anatomical characterization of origin, course, and termination, CMR provides exact quantification of shunt via phase velocity mapping and ejection fraction with global and regional myocardial function to create a road map for the interventionalist or surgeon guiding therapeutic approaches. A CMR perfusion scan can be added to assess the coronary steal phenomenon.

In our patients, CMR was able to demonstrate origin, epicardial course, and termination of the coronary fistula with respect to the coronary bed and the surrounding anatomy. In both cases the T_1 weighted axial spin echo images best delineated the origin and the entrance of the coronary vessels into the main pulmonary artery. The FIESTA cine sequences confirmed these findings. Also, the infundibular spin echo view was crucial to visualize the entry point in relation to the pulmonic valve plane. Multiple oblique imaging planes were complimentary to visualize and confirm the CAF. Phase velocity mapping was performed to accurately analyze stroke volume and quantitate shunt fraction. Although both cases exhibit left to right shunts, on CMR imaging, the turbulence was not noticed at the entry point of the fistulae suggesting near equalization of pressures or minimal shunt. Time of imaging for each patient was under 45 minutes and neither case required gadolinium.

We describe a fairly simple scanning approach to image patients with coronary anomalies. This approach can be used by centers currently not equipped with commercial coronary imaging platforms on CMR. After obtaining scout images, multislice axial T1-weighted images are obtained to localize coronary anatomy. Thereafter, breathold T1-weighted images with slice thickness of 6 mm and 0–3 mm overlap are acquired through the region of interest to further clarify the anatomy. Appropriate T1-weighted double oblique imaging planes such as right ventricular outflow tract for fistulae draining into the MPA or RV should be obtained. Similarly, very thin slices with overlap must be acquired on FIESTA images to localize and define the coronary tree. Phase velocity images are acquired by planning slices perpendicular to the aortic valve and pulmonic valve to obtain the right and left side stroke volume. In such scenarios meticulous care should be taken to position the slice distal to the termination of the CAF to accurately calculate the QP:QS.

5. Conclusion

In conclusion, we propose that CMR should be considered as an adjunct non-invasive diagnostic modality with coronary angiography to define the spatial localization, anatomy, and hemodynamic significance of coronary fistulae.

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